

Pancreas and Spleen

Pancreas

- Congenital variations
 - Ectopic pancreatic tissue
 - ▶ Pancreatic tissue may be functional, but patient is asymptomatic
 - ▶ Patient may have Meckel's diverticulum or intestinal duplications
 - ▶ Patient maintains exocrine function
 - ▶ Symptoms: bleeding and inflammation
 - ▶ Treatment: local resection if symptomatic
 - Annular pancreas
 - ▶ Etiology: pancreatic tissue circumferentially surrounds the second portion of the duodenum at the region of the sphincter of Oddi
 - ▶ Parenchymal and ductal structures are usually normal
 - ▶ Symptoms
 - ▷ Associated with atresia or stenosis of the underlying duodenum
 - ▷ Vomiting (may or may not be bilious) in newborns
 - ▶ Diagnosis
 - ▷ Antenatal polyhydramnios
 - ▷ Prenatal ultrasound and postnatal abdominal radiograph show "double bubble"
 - ▶ Treatment
 - ▷ Asymptomatic cases do not require treatment
 - ▷ Obstructive symptoms
 - Utilize right upper-quadrant transverse incision
 - Perform side-to-side duodenoduodenostomy (transverse incision in proximal duodenum, longitudinal incision in distal duodenum) as in correction of duodenal atresia
 - Duodenojejunostomy (alternate method)
 - Avoid gastrojejunostomy

- Pancreatic trauma
 - Etiology: blunt trauma (especially associated with bicycle or motorcycle handlebars; body may be transected when crushed over the spine)
 - Diagnosis
 - ▶ Check for persistent elevation of amylase; use magnetic resonance cholangiopancreatography (MRCP), or endoscopic retrograde cholangiopancreatography (ERCP) to rule out ductal injury (computed tomography [CT] scan may be normal)
 - ▶ Perform focused assessment with sonography for trauma (FAST) examination
 - ▶ Perform a CT scan (definitive study) with oral and intravenous (IV) contrast
 - Nonoperative management
 - ▶ Indications
 - ▷ Patient is hemodynamically stable
 - ▷ Transfusion requirement is < 50% of estimated blood volume (ie, < 40 mL/kg)
 - ▷ No transection of the pancreatic duct
 - ▶ Management
 - ▷ Admit to floor bed
 - ▷ Place patient under the care of a qualified pediatric surgeon
 - ▷ Ensure operating room and personnel are immediately available
 - ▷ Check serial hematocrit levels every 4 hours
 - ▷ Ensure there is cross-matched blood in the blood bank
 - ▷ Put patient on bed rest for 48 hours
 - ▷ Restrict patient's physical activity for 1 month
 - ▷ Perform ultrasound based upon clinical course and for follow up
 - Operative management
 - ▶ Injury to the body of the pancreas or pancreatic duct requires drainage; even if a ductal injury is not identified, it should be presumed and drained
 - ▶ Resect clearly nonviable pancreatic body or tail tissue
 - ▶ Treat transection or near-transection of the pancreatic duct

- ▷ Oversew or staple the distal end of the proximal pancreas
- ▷ Oversew or staple the proximal end of the distal segment and leave the entire distal segment in situ; resect the distal segment
- ▷ Perform distal pancreatectomy (usually with splenectomy) if injury is in tail
- ▷ Drain distal segment of pancreas by Roux-en-Y anastomosis to the jejunum
- Pancreatitis
 - Alcohol and biliary tract disease, common causes of pancreatitis in adults, are uncommon in children
 - Pathophysiology
 - ▶ Autodigestion (autolysis)
 - ▶ Exocrine secretions are stored in inactive forms in protective zymogens
 - ▶ Alkaline pH of exocrine glands and protease inhibitors assures protection
 - ▶ Pancreatic injury (trauma, obstruction, inflammation) can rupture protective membranes within the gland and activate digestive enzymes, causing autodigestion
 - Symptoms
 - ▶ Abdominal pain
 - ▶ Increased amylase
 - Diagnosis
 - ▶ Ratio of amylase to creatinine clearance > 6% indicates pancreatitis
$$\frac{\text{urine amylase} \times \text{serum creatinine} \times 100}{\text{serum amylase} \times \text{urine creatinine}}$$
 - ▶ Laboratory tests: serum amylase, lipase
 - ▶ Imaging methods: ultrasound, MRCP, ERCP (contraindicated in acute pancreatitis)
 - Etiology
 - ▶ The most common cause in children is blunt abdominal trauma
 - ▶ The most common cause of nontraumatic pancreatitis is idiopathic

- ▶ Pancreatitis can also be caused by medications (eg, steroids, chemotherapy), infections, especially viral infections (eg, mumps), cystic fibrosis, underlying biliary tract disease, hemolytic disease resulting in stones (eg, spherocytosis, sickle cell disease), or cholelithiasis
- Treatment
 - ▶ Medical
 - ▷ Stop offending medications
 - ▷ Control infection
 - ▷ Provide gut rest; insert nasogastric tube if patient is vomiting, administer total parenteral nutrition (TPN), and give analgesics (eg, meperidine, **not** morphine), volume replacement, antibiotics, H2-blockers, or somatostatin
 - ▷ Monitor arterial blood gasses for impending respiratory failure
 - ▷ Monitor lactate dehydrogenase and serum glutamic oxaloacetic transaminase, which indicate tissue necrosis
 - ▷ Monitor serum calcium for hypocalcemia (due to saponification); prescribe calcium gluconate as indicated
 - ▷ Monitor serum glucose for hyperglycemia; prescribe insulin as indicated
 - ▷ Monitor hematocrit for hemoconcentration or bleeding
 - ▶ Surgical
 - ▷ Debride obviously necrotic tissue
 - ▷ Drain (especially fluid in lesser sac)
 - ▷ Perform cholecystectomy for stones after resolution of the acute episode
- Pseudocysts
 - Trauma is the most common etiology in children
 - Cyst walls are composed of inflammatory tissue (**not** epithelium) and are usually located in the lesser sac
 - Symptoms: pain, compression or erosion of surrounding organs, secondary infection, hemorrhage, and perforation
 - Diagnosis
 - ▶ Upper gastrointestinal series will show anterior and superior displacement of stomach and downward displacement of colon

- ▶ CT or ultrasound may also be useful in diagnosis
- Treatment
 - ▶ Medical
 - ▷ 50% may resolve spontaneously in 3–4 weeks
 - ▷ Perform percutaneous drainage
 - ▷ Rest intestinal tract
 - ▷ Provide TPN
 - ▶ Surgical
 - ▷ Surgery is indicated in the case of persistence (> 6 wk) or recurrence, and when pseudocyst communicates with a major duct
 - ▷ Provide external drainage (if infected)
 - ▷ Provide internal drainage by Roux-en-Y cystenterostomy (unless infected)
 - ▷ Perform distal pancreatectomy if pseudocyst is in tail
 - ▷ Perform cystogastrostomy (open or endoscopic) if pseudocyst is in the pancreatic body or adherent to the stomach (bleeding after operation is a common complication resulting from stomach acid)
 - ▷ Perform Roux-en-Y cystjejunostomy if pseudocyst is located at the head of the pancreas or not adherent to stomach

Spleen

- Physiology
 - The thymus is the primary lymphatic organ during intra-uterine life
 - The spleen is the major site of hematopoiesis from birth to 6 months of age, when hematopoiesis is taken over by the bone marrow
 - The spleen produces immunoglobulin M (IgM) antibodies against encapsulated bacteria (eg, pneumococcus, hemophilus, meningococcus), and tuftsin and properdin, which enhance phagocytosis and stimulate production
 - Humeral immunity in neonates is transferred through the placenta (except IgM)
- Congenital anomalies
 - Asplenia (Ivemark syndrome): complete absence of the spleen
 - ▶ Also called asplenia syndrome or heterotaxia

- ▶ Can be associated with situs inversus (with or without malrotation or volvulus), cardiac anomalies (which are associated with a high mortality rate), and three-lobed lungs
- ▶ Symptoms
 - ▷ Congenital cyanotic heart disease, which results in cyanosis
 - ▷ Shortness of breath
 - ▷ Congestive heart failure
- ▶ Diagnosis
 - ▷ Howell-Jolly bodies will be evident in a peripheral smear
 - ▷ Perform a spleen scan or ultrasound
- Polysplenia
 - ▶ Multilobed spleen with 2–9 equal portions
 - ▶ Associated with situs inversus, cardiac anomalies, and biliary atresia
 - ▶ Patient exhibits normal splenic function
- Accessory spleen
 - ▶ Small nodules of splenic tissue apart from a normal-sized spleen
 - ▶ These are most commonly located in the splenic hilum (gastrosplenic ligament)
 - ▶ Nodules must be removed when a splenectomy is performed for hypersplenia to prevent persistent hematologic disease
- Cysts
 - ▶ Congenital cysts are rare
 - ▶ Epidermoid cysts are the most common type in pediatric patients
 - ▷ Symptoms: hemorrhage, left upper quadrant pain, infection
 - ▶ Echinococcal cysts are the most common type worldwide
 - ▶ Differential diagnosis: history, serology, scan
 - ▶ Diagnosis: ultrasound, spleen scan, CT scan
 - ▶ Treatment: perform partial cyst wall resection/marsupialization, rather than hemisplenectomy
- Trauma

- Diagnosis (see Pancreatic trauma)
- Nonoperative management (see Pancreatic trauma); patients who cannot be observed continuously in an intensive care unit (ICU) or who will be evacuated through multiple levels of care are **not** candidates for conservative management
- Operative management
 - ▶ Indications for operation
 - ▷ Patient requires transfusions of > 50% blood volume (~ 40 cc/kg)
 - ▷ Patient exhibits continued hypotension or evidence of continued hemorrhage
 - ▷ There is evidence of associated significant injuries
 - ▷ A patient with abdominal distension is in shock (immediate laparotomy is necessary)
 - ▶ Treatment
 - ▷ Splenorrhaphy is often possible
 - ▷ Perform splenectomy in an unstable patient with multiple injuries, or one who will not be continuously observed in an ICU setting or who will be transferred through multiple levels of medical care
 - ▷ Provide immunizations to protect against pneumococcus and vaccines to protect against *Haemophilus influenzae* and *Neisseria meningitidis*; give penicillin prophylaxis until the patient reaches 18 years of age
- Inflammation
 - Acute inflammatory splenomegaly
 - ▶ Etiology: infection is the most common cause
 - ▶ Treatment: treat the source of infection (do not perform splenectomy)
 - Abscess
 - ▶ Gram-negative anaerobic *Staphylococcus* is the most common cause
 - ▷ Hemoglobinopathy is associated with *Salmonella*
 - ▷ Leukemia is associated with *Candida*
 - ▶ Trauma
 - ▶ Infarction
 - ▶ Bacteremia
 - ▶ Immunosuppression

- ▶ Symptoms: fever, left upper-quadrant tenderness, and left shoulder pain
- ▶ Diagnosis: CT scan (best modality)
- ▶ Treatment
 - ▷ IV antibiotics
 - ▷ Percutaneous aspiration and drainage for large abscesses
 - ▷ Splenectomy (in the case of persistent infection)
- Hematologic disorders
 - Red blood cells (RBCs)
 - ▶ Hereditary spherocytosis
 - ▷ Autosomal dominant
 - ▷ Etiology: membrane is abnormal, which prevents RBCs from assuming discoid shape; deformity results in small, round, fragile RBCs
 - ▷ Symptoms: anemia, jaundice, splenomegaly, gallstones (gallstones occur in 75% of these patients)
 - ▷ Laboratory indicators: anemia, increased bilirubin, increased reticulocyte count, spherocytes, increased osmotic fragility
 - ▷ Treatment: splenectomy (deferred until age 3–4 years; most common indication) or cholecystectomy if stones present on preoperative ultrasound
 - ▷ If hemolysis recurs or if no Howell-Jolly bodies are seen in peripheral smear, suspect a residual accessory spleen
 - ▶ Elliptocytosis: rarely associated with RBC destruction sufficient to require splenectomy
 - ▶ Sickle cell anemia
 - ▷ Homozygous sickle cell gene produces severe, chronic, hemolytic anemia
 - ▷ Hemoglobin levels in individuals with sickle cell anemia are as follows:
 - HgB S: 90%
 - HgB F: 5%
 - HgB A2: normal
 - HgB A: absent
 - ▷ Sickling results in occlusion of small vessels (“sickle cell crisis”)

- ▷ Etiology: infection, dehydration, hypoxia, acidosis
 - ▷ Vasooclusive crisis is a potential complication characterized by pain and swelling in the hands and feet (“hand / foot syndrome”), acute abdominal pain, pulmonary infarction
 - ▷ Sequestration results in acute trapping of RBCs in the spleen, which leads to anemia, hypotension, and splenomegaly (if recurrent severe episodes [resulting in shock] occur, splenectomy is indicated after pneumococcal vaccine polyvalent and *H influenzae* type B [Hib] vaccine)
 - ▷ Eventually the spleen becomes small, fibrotic, and infarcted (functional asplenia); most common infections are pneumococcal and *Salmonella osteomyelitis*
 - ▷ Laboratory indicators:
 - HgB: 6–8 g/dL
 - Smear will show sickle, target, and nucleated RBCs; Howell-Jolly bodies; 5%–15% reticulocytes, increased white blood count, and increased platelets
 - Liver function tests: abnormal
 - Diagnosis: hemoglobin electrophoresis
 - ▷ Treatment
 - Hydration
 - Packed red blood cell (PRBC) transfusion to decrease HgB S to < 40% (20 cc/kg type-specific Rh[–] PRBCs) from usual 90%
 - Analgesics
- White blood cells
 - ▶ Leukemia
 - ▷ Splenomegaly secondary to leukemic infiltrate is the most common cause of splenic rupture
 - ▷ Splenectomy is not indicated
- Platelets
 - ▶ Idiopathic thrombocytopenic purpura (ITP)
 - ▷ Etiology: antiplatelet antibodies attach to platelets, making the platelets more susceptible to destruction in the spleen
 - ▷ History: patient will have had antecedent nonspecific viral illness

- ▷ Spleen is normal-sized, platelets are normal shape
- ▷ Symptoms: usually occurs in children (especially females) < 10 years old, a few weeks after a mild viral illness
- ▷ Physical: petechiae, bruising, bleeding (worst if purpura is in the central nervous system [CNS])
- ▷ Treatment: 75% go into spontaneous remission
- ▷ Treatment for chronic or persistent ITP
 - Steroids for 1–3 weeks in patients with persistent thrombocytopenia. Failure to respond to steroids may result in chronic ITP; patients most likely to respond to splenectomy are those in whom preoperative steroids have increased the platelet count (administer 100 mg hydrocortisone IV intraoperatively)
 - IV gammaglobulin (very expensive) is indicated if steroids fail
 - Anti-D immune globulin may be administered in some cases
 - Immunoglobulin G can be given to block platelet-antibody complex, resulting in a normal platelet count in 3–4 days after administration
- ▷ Indications for splenectomy (see Splenectomy) include an acute bleeding episode (especially in the CNS or intraabdominal space; this requires an emergency splenectomy)
 - Relapse is possible following steroids
 - Steroids may also result in persistent platelets < 10,000
 - Splenectomy is curative in > 90% of patients with ITP; however, if accessory splenic tissue is missed, symptoms will recur
- ▷ If platelet count is < 50,000, platelet transfusion should be given only until the splenic artery is ligated, at which time the platelet count will start to rise
- ▷ If postoperative thrombocytosis occurs (platelets number > 1,000,000), prescribe aspirin
- Hypersplenism
 - ▶ Pancytopenia (decreased white blood cells, RBCs, or

- platelets) may be primary or secondary to portal hypertension, inflammatory diseases, storage disease, chronic hemolytic disease, myeloproliferative disorder, or neoplastic disease
- ▶ Hypersplenism is associated with sickle cell crisis
 - ▶ Treat primary hypersplenism with splenectomy; however, splenectomy is not indicated for secondary hypersplenism resulting from Hodgkin disease, sarcoid, leukemia, and portal hypertension
 - Wandering (ectopic) spleen
 - Symptoms: when spleen is attached only by hilar vessels, torsion may occur, leading to abdominal pain
 - Most common in male infants
 - Diagnosis: ultrasound
 - Treatment: splenectomy if the spleen is not infarcted
 - Splenectomy
 - Most common indication for splenectomy is ITP
 - Preoperative preparation
 - ▶ Immunize patient with polyvalent capsular polysaccharide antigens of pneumococcal vaccine ideally administered > 3 weeks before elective splenectomy, and Hib vaccine
 - ▷ Pneumococcal vaccine is only effective against 80% of organisms (may be less effective in children < 2 y old) and provides protection for children 4–5 years old
 - ▶ Administer prophylactic antibiotics before, during, and after operation, and regularly until patient is 18 years old
 - ▷ Use ampicillin in children < 10 years old
 - ▷ Use penicillin if patient is > 10 years old
 - ▶ Give an intraoperative stress dose of steroids (100 mg hydrocortisone IV) if patient was treated with steroids immediately before the operation
 - Operative procedures
 - ▶ Place a nasogastric tube to prevent gastric distension and dislodgement of ties on the short gastric vessels
 - ▶ Make a laparoscopic, upper midline, or left subcostal incision
 - ▶ Mobilize by incising posterior, lateral peritoneal reflection

- ▶ Divide short gastric vessels
- ▶ Ligate and divide splenic artery
- ▶ Avoid injury to the tail of the pancreas
- ▶ Search for accessory spleens in gastrosplenic ligament
- ▶ All splenic tissue must be removed for hematologic reasons or the disorder will recur
- Postoperative considerations
 - ▶ The most serious postoperative complication is overwhelming postsplenectomy infection (see below), which is associated with a 50% rate of mortality, especially in children < 2 years old
 - ▶ Laboratory tests
 - ▷ Peripheral blood smear will show the following cytoplasmic inclusions:
 - Heinz bodies
 - Howell-Jolly bodies
 - Siderocytes
 - ▷ Increased white blood cell count
 - ▶ Thrombocytosis
 - ▷ A platelet count < 1,000,000 requires no treatment
 - ▷ Platelet count may be > 1,000,000 10 days after operation; treat with aspirin (80 mg/day)
 - ▷ Thrombotic complications (eg, portal vein thrombosis, diagnosed by ultrasound) are rare
 - ▶ Overwhelming postsplenectomy infection
 - ▷ Risk
 - Greatest in infancy, decreases with age
 - Twice as great in children < 2 years old
 - Least when splenectomy is done for trauma
 - Greatest when splenectomy is done for thalassemia and other hematological indications
 - Overall incidence ~ 5%
 - ▷ 80% occur within 2 years of splenectomy
 - ▷ Symptoms (extremely rapid in onset and progression)
 - Nausea and vomiting
 - Confusion
 - Seizures
 - Shock

- Disseminated intravascular coagulation
- Coma
- Death (50%)
- ▷ Pneumococcus is the most common organism responsible, **not** hemophilus
- ▷ Prevention: polyvalent pneumococcal vaccine

